Syndrome of Left Ventricular-Right Atrial Shunt

Successful Surgical Repair of Defect in Five Cases, with Observation of Bradycardia on Closure *

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THE VARIATION of a membranous ventricular septal defect which has its major flow into the right atrium has been recognized for many years. More recently the opportunity which advances in intracardiac surgery offer has made it possible to close such defects with relatively low mortality rate. This makes it desirable to study the variations in clinical and laboratory observations which facilitate the recognition in advance of the exact location of the shunt. It is the purpose of this paper to describe the essential clinical features of five instances of left ventricular-right atrial shunt. All were successfully closed with the aid of extracorporeal circulation. It was of interest to observe that upon temporary closure of the defect manually, with an intact circulation, the systemic blood pressure rose and the pulse slowed in an identical manner as has been observed to take place upon closure of a peripheral arteriovenous fistula or a patent ductus arteriosus.

Although such lesions have been described for many years, only five patients have been reported who have had a complete clinical study including cardiac catheterization and in whom surgical repair of the lesion has been attempted.[9, 10, 14, 20] One of these patients survived.[9] An earlier case report called attention to the cardiac catheterization findings that one would expect in the presence of the lesion, although in this case this study was not done.[18]

In addition to there being an opening, which is essentially between the left ventricle and right atrium, there is usually some defect in the septal leaflet of the tricuspid valve adjacent to the margin of the shunt. This further allows the escape of blood into the right atrium.

Material and Methods

Phonocardiograms were recorded using a Sanborn Twin Beam Cardiette at paper speeds of 75 mm. per second. Murmurs were graded from 1 to 4 according to their loudness. Catheterization studies were performed in the usual manner and all blood samples were analyzed using the Van Slyke technic. Blood flows were calculated using the Fick principle and assuming a normal oxygen consumption for each patient. Pulmonary venous blood was assumed to be 95 per cent saturated. Inferior and superior vena caval blood oxygen contents were averaged to obtain the peripheral arteriovenous difference. If only a superior vena cava sample was obtained this value was used. In each patient the ratio of pulmonary blood flow to peripheral blood flow was

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TABLE 1

D. L. 11–12–53		
Oxygen content ml./100 ml. blood		
SVC	11.1	
	11.5	
IVC	11.8	Pressure measurements mm. Hg above mid-chest
RA	13.7	RA 3 (mean)
RA	13.8	RV 11/3
RV	14.2	PA 7 (mean)
RV	15.4	Estimated pulmonary flow 3.7 L./min./m.^{2}
PA	14.8	Estimated peripheral flow 2.1 L./min./m.^{2}
O_2 capacity	19.8	Ratio pul. flow/per. flow 1.8
PCV	42	

calculated. One catheterization study (Case 3) was performed at another hospital.

Case Reports

Case 1. D. L. This 15-year-old schoolboy was first seen in 1952, at the age of 10, for evaluation of a heart murmur which had been present since birth. He was normally active but had noted that he became tired easily. Physical examination revealed a well-developed boy in no distress. The blood pressure was 120/80. There was no evidence of cardiac enlargement. In the 4th interspace along the left sternal margin a grade 4/4 systolic murmur was present as well as a faint thrill. The murmur had a curious terminal musical component suggestive of a "gull cry" murmur. The murmur was not loud at the pulmonic area. The second sound in this area was accentuated. A clinical diagnosis of an interventricular septal defect was made and cardiac catheterization studies were performed on November 12, 1953. The results are summarized in Table 1. It was concluded that an atrial septal defect was present but that an associated small interventricular septal defect could not be excluded.

In August 1956 the patient was seen again because of an increase in his fatigue and the presence of mild exertional dyspnea. The physical findings were essentially unchanged. An electrocardiogram revealed a slight delay in right ventricular conduction with a QRS duration of 0.09 second (Fig. 2). A phonocardiogram recorded from the left sternal border at the 4th interspace revealed a holosystolic murmur with a late musical component (Fig. 1A). Aortic closure preceded pulmonic closure during expiration of 0.05 second. Roentgen studies of the heart and lungs revealed slight enlargement of the right ventricle and pulmonary artery without an increase in the pulmonary vascular markings. The aortic knob appeared small (Fig. 3). In addition there was anterior wedging of the 8th thoracic vertebral body probably due to juvenile epiphystitis.

Cardiac catheterization studies were performed for the second time on April 24, 1957, and the results are summarized in Table 2. The data were thought to be compatible with a small shunt at the atrial level and possibly a mild pulmonic stenosis.

Operation was performed on April 25, 1957, with the aid of extracorporeal circulation. A small septal defect was found penetrating the septal leaflet of the tricuspid valve near its margin of insertion (Fig. 4A). The heart was stopped and the defect repaired under direct vision. After closure of the atrial incision ventricular fibrillation appeared, but this was effectively reversed by cardiac massage and electrical defibrillation. The postoperative course was uneventful. The patient was seen four months after surgery. He was asymptomatic even during strenuous activity. The murmur had diminished in intensity and the thrill previously noted was absent. At the pulmonic area the phonocardiogram demonstrated a faint 2/4 mid-systolic ejection murmur. The sound of pulmonic closure was accentuated and followed the sound of aortic closure by 0.04 second during expiration.

Comment: The reason for the apparent decrease in magnitude of the left-to-right shunt between the two catheterization studies is not clear. Both studies were performed using sedation and local anesthesia only, but apprehension may have been greater during the initial study and may be responsible for the greater shunt at that time. The location of the murmur and thrill

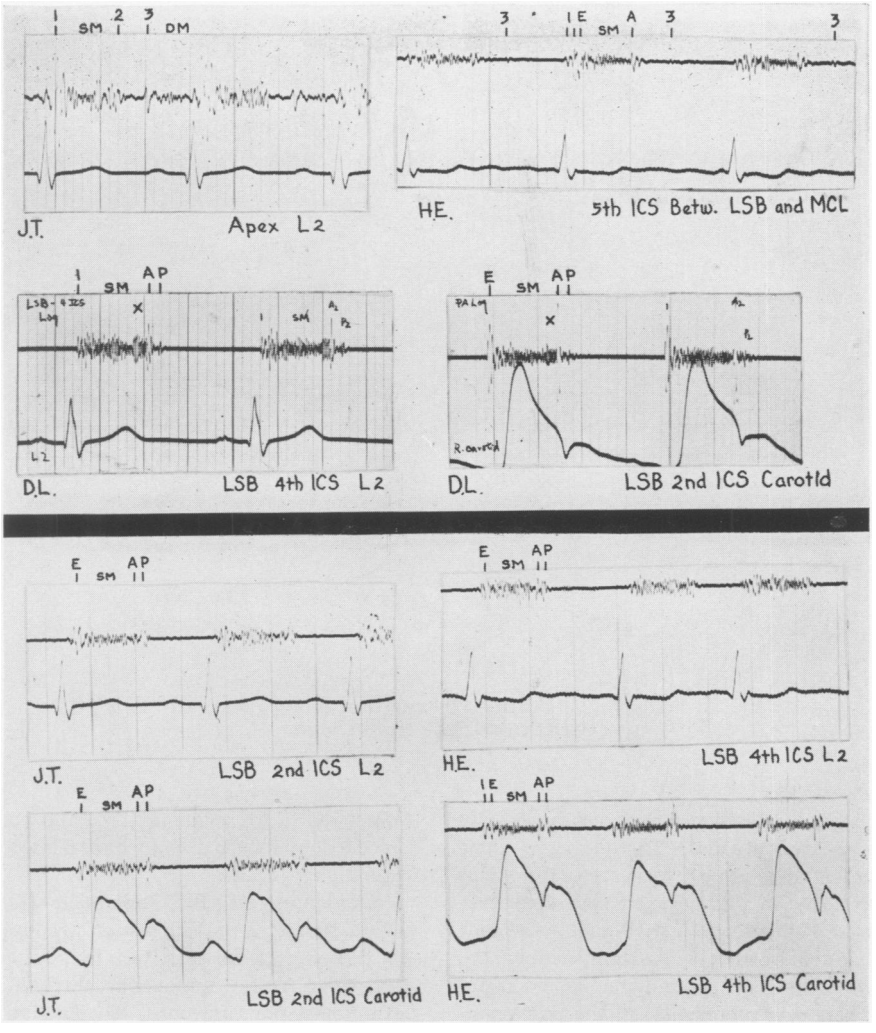


FIG. 1a (upper) and b (lower). Preoperative phonocardiograms of three patients with left ventricular-right atrial shunts. The following abbreviations have been used: 1 = first sound, SM = systolic murmur, 2 = second heart sound, 3 = third heart sound, DM = diastolic murmur, E = ejection sound, A = aortic valve closure, P = pulmonic valve closure.

at the 4th interspace and the holosystolic character of the murmur as demonstrated by the phonocardiogram are compatible with a ventricular septal defect.

Case 2. J. T. This girl was seen at Stanford University Hospital for the first time in August 1948, at the age of 18 months, because of the discovery by her mother of a heart murmur. The child had been noted to have poor growth and became tired quite easily. There were bilateral clubbed feet; no cyanosis had been observed. Two subsequent episodes of congestive failure occurred in association with acute infectious disease. In July 1953, cardiac catheterization was performed (Table 3) and a diagnosis of atrial septal defect was made. In January 1957, the patient was admitted to hospital where physical examination revealed a very thin girl of gracile habitus. There was a slight sternal protrusion and a distinct right ventricular heave was felt, and a systolic thrill could be palpated to the left of the sternum. The blood pressure was 110/80. There was a grade 4/4 harsh holosystolic murmur over the entire precordium, loudest at the left sternal margin at the 4th inter-

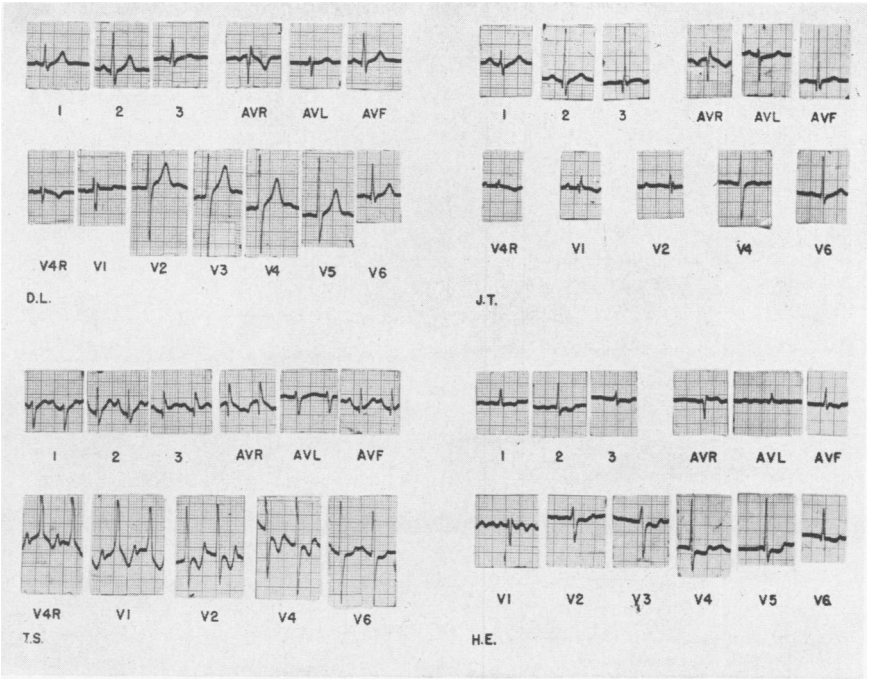


FIG. 2. Preoperative electrocardiograms of four patients with left ventricular-right atrial shunts.

space. The second pulmonic sound was moderately split, but not accentuated. There was a faint diastolic rumble at the apex. An electrocardiogram revealed a delay in right ventricular activation (Fig. 2) and roentgenological studies showed moderate cardiac enlargement involving mainly the right atrium, right ventricle and pulmonary artery (Fig. 3). There was prominence of the pulmonary vascular markings. In addition to the above-mentioned murmurs phonocardiograms showed an ejection sound in the pulmonic area and aortic closure preceded pulmonic closure by 0.04 second during expiration (Fig. 1B).

On January 18, 1957, operation was performed with a preoperative diagnosis of atrial septal defect. On exposing the heart is was apparent immediately that there was a prominent thrill in the right atrium near the atrioventricular juncture. The right atrium was quite distended, red and bulged with ventricular systole. Palpation through an atriotomy

TABLE 2

D. L. 4–24–57			
Oxygen content ml./100 ml. blood			
SVC	13.5		
		14.0	PA 14.8
IVC	14.4		
			14.5
RA	14.9		
	14.5		BA 17.8 = 93%
	14.4		O_2 capacity 19.2
	14.3		Estimated pulmonary flow 4.0 L./min./m.^{2}
RV	14.6		Estimated peripheral flow 3.7 L./min./m.^{2}
	14.8		Ratio pul. flow/per. flow 1.1
	14.8		Pressure measurements mm. Hg above mid-chest
			RV 25/5 PA 19/9 BA 125/77

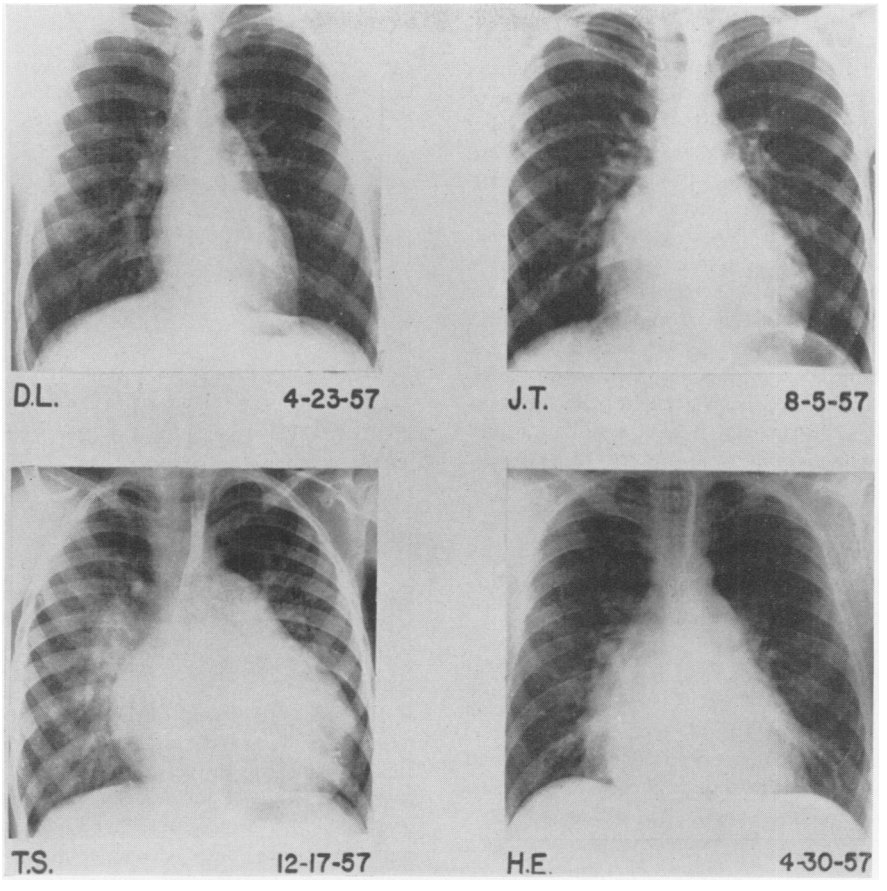


FIG. 3. Preoperative x-rays of four patients with left ventricular-right atrial shunts.

revealed a defect in the upper margin of the septal leaflet of the tricuspid valve (Fig. 4A). This defect could be closed easily with the index finger, eliminating the thrill, slowing the heart and raising the systemic blood pressure (Fig. 5). It was decided that this lesion could best be corrected using extracorporeal circulation. The operation was therefore terminated and the patient allowed to recover.

On August 8, 1957, operation was again per-

TABLE 3

J. T. 7–14–53		
Oxygen content ml./100 ml. blood		
SVC	11.1	
	10.7	Estimated pulmonary flow L./min./m.^{2} 5.7
IVC	10.3	
		Estimated peripheral flow L./min./m.^{2} 3.3
RA	13.5	
		Ratio pul. flow/per. flow = 1.7
RA	12.2	
RV	13.2	Pressures mm. Hg above mid-chest
FA	15.5–92%	RA 2 (mean)
O_2 capacity	16.8	RV 41/1
PCV	44 (11.22–53)	FA 86/51

formed with the aid of extracorporeal circulation. The thorax was opened through a median sternotomy and the previously mentioned bradycardia and rise in systemic pressure on manual closure of the defect was again recorded (Fig. 5). Utilizing elective cardiac arrest with potassium [5] and approaching the defect (Fig. 4A) through an atriotomy the 12 mm. opening between the left ventricle and right atrium was closed with interrupted sutures. There was also a defect in the upper margin of the septal leaflet of the tricuspid valve directly overlying the opening. The atrium was repaired in the usual manner and the heart restarted in normal sinus rhythm. The postoperative course was satisfactory and there has been a remarkable improvement in physical capacity. A faint systolic murmur is still present over the mid-precordium.

Comment: The physical findings were those of ventricular septal defect, although the catheterization data suggested interatrial septal defect.

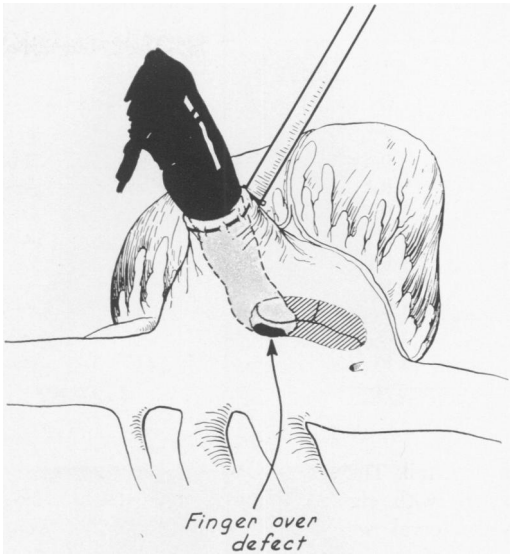


FIG. 5. Drawing showing manual closure of defect with resulting rise in systemic blood pressure.

Case 3. T. S. A 5-year-old boy was referred to Stanford University Hospital on December 16, 1957, for surgical repair of a congenital cardiac lesion. A heart murmur and transient cyanosis had been noted at birth. Following this there had been frequent attacks of pneumonitis and at 6 weeks congestive failure had appeared with a heart rate of 174 and enlargement of the liver. The child had received digitalis and diuretics since that time. The clinical course up to the present hospital entry had been characterized by numerous episodes of congestive failure usually associated with respiratory tract infections and accompanied by dyspnea, tachycardia, hepatic enlargement and occasional attacks of unconsciousness. Cyanosis had subsequently never been noted. Diagnostic studies had been performed at two other hospitals in 1956 with the following results: Pulmonary artery pressure 54/24; right ventricular pressure 54/8 and right atrial pressure 14/7 mm. Hg. A left-to-right shunt had been demonstrated in the right atrium and right ventricle. Oximetric studies had revealed an arterial saturation of 95 per cent at rest and 94 per cent during exercise.

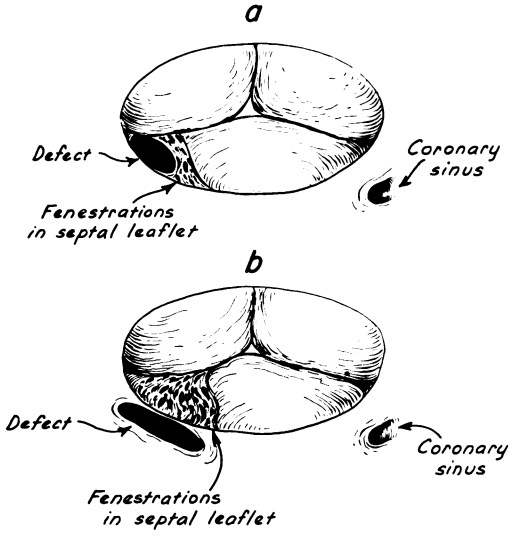


FIG. 4. Drawing showing the essential aspects of left ventricular-right atrial shunt. a) Type of defect in 4 out of the 5 patients. b) Type of defect in Case 3.

Physical examination revealed a normally developed boy with a blood pressure of 98/60. A moderate pigeon-breast deformity was present. A right ventricular heave was noted and at the left sternal margin and in the 3rd and 4th interspaces a grade 4/4 systolic murmur with an accompanying thrill was present. A faint grade 1/4 diastolic rumble was present at the apex. The second sound was accentuated and distinctly split at the pulmonic area. Rales were present at both lung bases, the liver edge was palpated 2 cm. below the costal margin and slight peri-orbital edema was noted. The electrocardiogram revealed a right axis deviation (150°), tall (3.5 mm.) peaked P waves in the right precordial leads, and the R wave in V_1 was

TABLE 4

T. S. 11-19-57

Oxygen content ml./100 ml. blood

SVC	9.4	Estimated pulmonary flow L./min./m.^{2} 12.5
RA	12.6	Ratio pul. flow/per. flow = 3.5
RA	13.1	Pressure measurements mm. Hg above mid-chest
RV	13.7	RA 5 (mean)
RV	14.3	RV 60/0
PA	13.2	PA 60/25
PA	13.2	Aorta 90/50
Aorta	14.3	The catheter was thought to pass from the right
O_2 capacity	15.4	ventricle through a ventricular septal defect into
PCV	34.5% (12-17-57)	the ascending aorta and the innominate artery

16 mm. tall. These data were thought to be compatible with right ventricular hypertrophy. The P-R interval was 0.20 second (Fig. 2). X-ray studies of the heart and lungs revealed a marked cardiac enlargement with enlargement of both atria and both ventricles. The pulmonary artery segment was prominent and the peripheral pulmonary vessels were congested (Fig. 3). Cardiac catheterization was performed on November 19, 1957 (Table 4). A diagnosis of a combined atrial and ventricular septal defect with moderate pulmonary hypertension and a left-to-right shunt was made.

On December 19, 1957, the thorax was opened through a median sternotomy. The heart was greatly enlarged, particularly the right ventricle and right atrium. The right atrium pulsated vigorously with each beat, was quite red, and there was a sharp thrill over the anterior aspect of the heart at the atrioventricular juncture. Palpation through a right atriotomy revealed a defect large enough to admit the index finger in the margin of the septal leaflet. Manual closure of the defect caused bradycardia and rise in systemic pressure. Utilizing total heart-lung bypass and elective cardiac arrest with potassium [5] the right atrium was opened. A defect measuring 2 cm. in diameter presented into the right atrium just at the margin of the septal leaflet of the tricuspid valve (Fig. 4B). Probing confirmed its communication with the left ventricle. There was some fenestration of the tricuspid valve in this area, and the margin of the defect showed quite a lot of turbulence fibrosis. Closure of the defect was done with interrupted silk sutures, and the fenestrations in the tricuspid valve were drawn together over the repair. When the atriotomy was closed and the heart restarted there was no further thrill, and a normal sinus rhythm was present. He had a good recovery and no longer has a murmur.

Comment: This patient had severe symptoms due to pulmonary congestion and there was doubt whether or not he would survive before surgery. Postoperatively he is remarkably improved.

Case 4. H. E. This 42-year-old clerk was referred to the Stanford University Hospital on April 29, 1957, for evaluation of a congenital cardiac lesion. Transient cyanosis had been noted at birth, as well as a heart murmur. No symptoms were experienced throughout childhood, but at the age of 20 palpitation and mild exertional dyspnea appeared. Eight years prior to entry these symptoms became more severe and frequent; nearly annual attacks of pneumonitis were experienced. Four months prior to entry the patient was unable to work because of exertional dyspnea, wheezing and palpitations, which limited his activity to two flights of stairs. He began to have frequent attacks of nocturnal dyspnea relieved by sitting up in a chair or lying prone. There had been no edema. There was no history of rheumatic fever. He had been receiving digitalis for several months prior to entry. Physical examination revealed a moderately obese, ill-looking male lying flat in bed. The blood pressure was 145/70 and the heart was totally irregular with an apical rate of 90. The neck veins were flat and there was no cyanosis. Cardiac dullness extended to the anterior axillary line in the 5th interspace, where the apex impulse was diffuse. A distinct right ventricular parasternal heave was present. At the 5th interspace along the left sternal border a grade 4/4 holosystolic murmur was heard and a systolic thrill was present. The murmur was well transmitted to the apex, but only faintly heard at the pulmonic area. The second sound at the pulmonic area was not accentuated and no abnormal splitting was noted. No gallop sounds were heard. Fine moist rales as well as

TABLE 5

H. E. 5-2-57		
Oxygen content ml./100 ml. blood		
SVC	10.9	Estimated pulmonary flow 3.7 L./min./m.^{2}
	11.1	
IVC	11.3	Estimated peripheral flow 2.1 L./min./m.^{2}
RA	14.2	Ratio pul. flow/per. flow 1.8
RA	14.1	
		Pressure measurements mm. Hg above mid-chest
RV	14.1	
RV	14.5	RA 6 (mean)
PA	14.4	RV 51/9
PA	13.9	PA 49/32
BA	17.3-92%	
O_2 capacity	19.0	PCV 51% (4-29-57)

transient expiratory and inspiratory wheezes were present at both lung bases. The liver was not enlarged and there was no edema. The arm-to-tongue circulation time (Decholin®) was 15 seconds. The vital capacity was 2.4 liters in nine seconds.

An electrocardiogram revealed atrial fibrillation and abnormal T waves (Fig. 2). Roentgen studies of the heart and lungs revealed enlargement of the right ventricle, both atria and the pulmonary artery. The peripheral pulmonary vessels were prominent and a hilar dance was noted at fluoroscopy (Fig. 3). The phonocardiogram revealed a holosystolic murmur at the sternal margin. A third heart sound was present at the apex. Pulmonic closure followed aortic closure by 0.04 second during expiration (Fig. 1A-1B).

A clinical diagnosis of an atypical atrial septal defect, possibly of the septum primum variety, was made and cardiac catheterization studies were performed on May 2, 1957 (Table 5). The data appeared to confirm the clinical impression and on October 31, 1957, the patient re-entered the hospital for surgical repair of the defect. The arm to tongue circulation time had increased to 22 seconds.

Operation was performed on November 5, 1957. The thorax was opened through a median sternotomy. Exposure of the heart revealed an enlarged right ventricle and a very large red right atrium which bulged vigorously with each ventricular systole. There was a sharp systolic thrill over the atrioventricular juncture. Palpation through an atriotomy revealed an opening about one centimeter in diameter, which when occluded caused bradycardia and eliminated the thrill. The patient was placed on total heart-lung bypass and a right atriotomy was performed. The defect was found to be in the lower corner of the tricuspid valve orifice, and communicated with the left ventricle (Fig. 4A). It was closed with an Ivalon® sponge and interrupted sutures. After repairing the atrium the thrill and the distension were no longer present.

The postoperative course was without incident and the patient was discharged from the hospital on November 22, 1957, at which time no murmur was audible.

Comment: This is a patient who had rather severe cardiac symptoms, whose physical signs were of a ventricular septal defect. Catheterization data suggested an atrial septal defect.

Case 5. T. A. This was a 17-year-old Eskimo girl who was known to have had a heart murmur since infancy. As she grew she noted occasional palpitation, increasing fatigability, and only occasional respiratory infections. On physical examination she was well developed and nourished. There was no evidence of cyanosis or clubbing. There was a loud systolic murmur over the entire precordium with maximum intensity in the 4th left interspace. The murmur was noted to extend across the sternum to the right rather markedly and there was a systolic thrill to the left of the sternum at the 3rd and 4th interspace. Roentgenograms revealed cardiac enlargement mainly involving the right ventricle; the pulmonary artery segment was prominent and showed marked pulsation. A phonocardiogram showed that the second sound was split by 0.05 second with the second component fainter than the first. A loud holosystolic murmur with a midsystolic accentuation was recorded at the lower left sternal margin. Apex phonocardiograms demonstrated a third heart sound occurring 0.18 second

TABLE 6

T. A. 2–17–58

Oxygen content ml./100 ml. blood

SVC	10.9	Pressure measurements mm. Hg above mid-chest
RA	9.9	RA 1 (mean)
RA	11.1	RV (low) 44/4
		RV (high) 24/4
RV	12.9	PA 24/6
RV	12.69	Estimated pulmonary flow 8.0 L./min./m.^{2}
PA	12.3	Estimated peripheral flow 3.8 L./min./m.^{2}
PA	11.8	Ratio pul. flow/per. flow 2.1
O_2 capacity	14.8	

after the aortic valve closure. Cardiac catheterization on February 17, 1958, was interpreted as showing a ventricular septal defect with a moderate left-to-right shunt and probably a mild infundibular stenosis (Table 6). The electrocardiogram was normal except for a P-R interval of 0.21 second.

Operation was performed on February 26, 1958, the thorax being opened through a median sternotomy. Upon exposure of the anterior surface of the heart the right atrioventricular juncture bulged with each systole and there was a sharp thrill at the site anteriorly. The right atrium was obviously under pressure. Palpation within the right atrium revealed an opening in the corner of the tricuspid valve with an obvious shunt into the right atrium (Fig. 4A). Manual closure of the defect produced bradycardia and a rise in systemic blood pressure.

Using total heart-lung bypass and elective cardiac arrest with potassium [5] exposure was obtained through a right atriotomy. The defect measured 12 mm. in diameter and was closed with a series of interrupted sutures. There was some absence of the upper margin of the septal leaflet near the defect and to prevent regurgitation through this corner of the tricuspid valve the entire leaflet at the corner was sutured over the repair. The heart was allowed to restart, which it promptly did, in normal sinus rhythm.

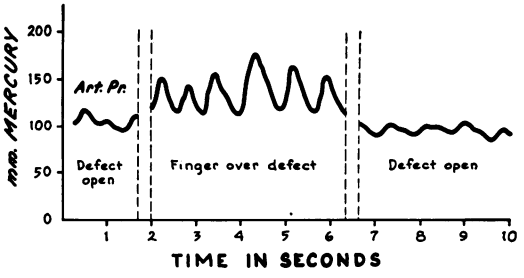
Postoperative recovery was entirely uneventful and there was no residual murmur.

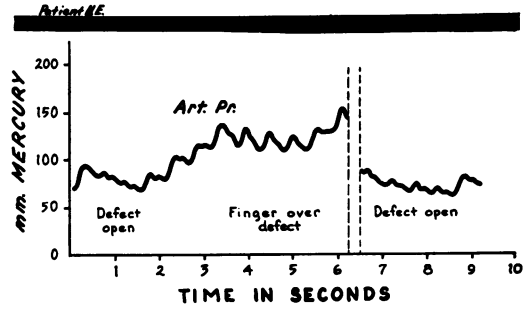
Comment: This was a left ventricular-right atrial shunt which on catheterization studies was interpreted to be a ventricular septal defect. It is difficult to understand why the blood sample taken low in the right atrium did not have a higher oxygen content, for it was obvious that a great deal of the shunt was into the atrium.

Discussion

The anatomic variations of the syndrome of left ventricular-right atrial shunt have been summarized by Perry, Burchell and Edwards.[18] Reports of other cases not mentioned by these authors have been made by several workers.[8, 15, 16, 19, 21, 22] Three general varieties of communication should be considered: (1) Fusion of the septal leaflet of the tricuspid valve to the edges of the ventricular septal defect associated with a perforation of the leaflet. The shunt occurs from the left ventricle directly into the right atrium. (2) A defect or cleft of the tricuspid valve leaflet at its point of attachment directly overlying the ventricular defect. The shunt occurs from the left ventricle to the right ventricle and, by regurgitation, into the right atrium. (3) A combination of these two lesions allowing a varying proportion of shunted blood to enter the right atrium and right ventricle. The lesions observed in the patients in this study are illustrated in Figure 4A & B.

Considerable attention was paid to such lesions by earlier workers since the tricuspid valve deformity was frequently associated with localized endocardial thickening which was thought to be evidence of fetal endocarditis and therefore suggested an inflammatory origin for the congenital defect.[7] It is quite possible that the endocardial thickening opposite the septal defect is not inflammatory in origin but represents





Patient J.T.

FIG. 6 (Upper). Drawing of systolic arterial blood pressure in Case 4 as recorded by an electromanometer connected to a small catheter in the left internal mammary artery. The average pulse rate just preceding the closure of the defect was 90 beats per minute; this was reduced to 60 beats per minute with the defect closed. It returned to 72 beats per minute wtih the defect open.

FIG. 7 (Lower). Drawing of systolic arterial blood pressure in Case 2 as recorded by an electromanometer connected to a small catheter in the left internal mammary artery. The average pulse rate just preceding the closure of the defect was 116 beats per minute and this was reduced to 100 beats per minute with the defect closed. It returned to 116 beats per minute with the defect open.

a jet lesion related to mechanical trauma of the endocardium due to turbulence as suggested by Edwards.[18]

The essential physiologic feature of the lesion consists of the shunt from the left ventricle to the right atrium. Because of the low mean pressure in the right atrium throughout the cardiac cycle a high pressure gradient is present across the defect throughout systole. Therefore a small orifice would allow a greater shunt to occur with this variety of communication than would occur with a similar sized orifice in the ordinary variety of ventricular septal defect, where the gradient would be reduced by the rise of pressure in the right ventricle during systole.

A distinctive feature of these lesions noted at surgery is the presence of a systolic expansion of the right atrium which is characteristically enlarged. Systolic atrial expansion is probably due to the fact that the shunt occurs largely during systole and that the high pressure left ventricle is ejecting into a right atrium closed at its outlet by the tensed tricuspid valve. Since the atrium is in communciation with the capacious venous reservoirs of the great veins its expansion occurs without much pressure change. Examination of pressure tracings from the right atrium therefore do not reveal a prominent pressure elevation during ventricular systole. Although the right atrium may be large in an atrial septal defect, no systolic expansion is usually seen with this lesion because the flow occurs throughout the cardiac cycle. This can readily be detected when the finger is placed in the defect, when a continuous flow will be felt.

Of unusual interest was the finding that on manual closure of the defect there was a bradycardia and rise in systemic blood pressure (Fig. 5–7). This phenomenon, previously described by Nicoladoni [17] and Branham [2] on closure of a peripheral arteriovenous fistula, has been thought to be due to the elimination of the reduced peripheral resistance at the fistula which allows the full output of the left ventricle to pass into the systemic circulation. This causes stimulation of the carotid sinus mechanism and the depressor nerves which then reflexly, through the vagus, slows the heart. It is believed that the effect shown here is similar physiologically to that observed on closure of an arteriovenous fistula, for the left ventricular-right atrial shunt is essentially an internal fistula. This phenomenon was observed in all of the five patients operated upon. Similar observa-

tions have been made upon closing experimental interventricular communications in the dog.[4]

It is pertinent to inquire why patients with ventricular-right atrial shunts usually have symptoms, especially dyspnea and pulmonary congestion, when patients with uncomplicated atrial septal defects with frequently even larger shunts may be asymptomatic. The probable explanation is that in the former lesion with an intact atrial septum, left atrial pressure and hence pulmonary capillary pressure may be elevated, thus producing pulmonary congestion and dyspnea. This is supported by the frequent finding of left atrial enlargement by x-ray. In an atrial septal defect left atrial pressure is normal and pulmonary venous congestion does not occur except in the rare instance of bi-ventricular failure. Left atrial enlargement is rarely observed by x-ray.[1]

The essential diagnostic feature of the left ventricular-right atrial shunt is the clinical picture of a ventricular septal defect in the presence of cardiac catheterization data indicating an atrial septal defect.

Auscultation will reveal the presence of a loud, harsh holosystolic murmur usually located at the left sternal margin in the 4th or 5th intercostal space. An associated thrill is usually present. Wide splitting of the second sound at the pulmonic area, usually found with an atrial septal defect, is absent.

At the apex a prominent third heart sound and a mid-diastolic rumble may be present. The systolic murmur of an atrial septal defect, on the other hand, is of variable intensity and consists of a mid-systolic injection murmur usually maximal at the pulmonic area. A third heart sound is only rarely present in an atrial septal defect and a diastolic murmur, when present, is usually located along the sternal margin.[11]

The electrocardiogram usually reveals evidence of a delay in right ventricular activation producing an incomplete right bundle branch pattern. A prolonged P-R interval (3 cases) and peaked P waves in the right precordial leads (2 cases) may be related to the right atrial dilatation and hypertrophy.[6] Tall peaked P waves (P pulmonale) are rarely found in the usual atrial septal defect. This abnormality was not present in any of 40 patients with proven uncomplicated atrial septal defects studied in this laboratory. Similar observations have been made by Barber.[1] A prolonged P-R interval, however, may occasionally be present in an atrial septal defect.[1]

The phonocardiogram provides a useful method of objectively examining the characteristics of the sound and murmurs and also enables one to differentiate this lesion from an atrial septal defect. The systolic murmur is loud, it begins with mitral valve closure, it lasts throughout systole and ends shortly after aortic valve closure. There is no mid-systolic accentuation. The aortic and pulmonic components of the second sound are not widely separated (0.03 to 0.05 second during expiration). A third heart sound and a diastolic rumble may be present at the apex. The murmur is probably due to increased mitral flow and is analogous to similar murmurs present in ordinary ventricular septal defects with increased pulmonary blood flow.[23]

The radiologic features of the lesion are essentially similar to those of a ventricular septal defect with a left-to-right shunt, with the added finding of distinct and sometimes marked right atrial enlargement which may form the convex border of the lower left portion of the cardiac shadow in posterior-anterior views. Enlargement of the right ventricle, pulmonary artery and congestion of the pulmonary vessels compatible with increased flow are present as well as distinct left atrial enlargement. Left ventricular enlargement is usually present but may be difficult to detect in the presence of right ventricular enlargement. The aortic knob is small. Left atrial and left ventricular enlargement are not found in uncomplicated atrial septal defects.

TABLE 7. *Summary of Essential Features of Eleven Patients with Left Ventricular-Right Atrial Shunts*

No.	Age	Sex	Symptoms 0–4+	Murmur Location Character Loudness	Thrill	ECG	X-ray Data RA LV 0–4+ LA PA Aorta RV Pul. Vasc. Engorgement	Catheterization Data Shunt Level	Pul. Flow/ Per. Flow	RV Press.	Size of Defect	Course	Reference
1	5 yrs.	F	0	Base, loud, harsh, systolic	Present	High voltage biphasic complexes in Leads 1 & 3 PR = 0.18 sec.	Slight cardiac enlargement PA prominent	Not catheterized			3 mm.	Died of S.B.E.	Edwards
2	6 mos.	M	0	LSB 3rd ICS mod. loud, harsh, systolic	?	RVH Inc. RBBB P pulmonale	RV +++ LV ? LA ++ PA +++ RA ++ Pul. vasc. engorgement	RA ? RV Cath. entered LA	1.5	37/6	5 mm. Small 5 mm. pat. foramen ovale also present	Died during surgery	Kjellberg
3	4 yrs.	M	++++	LSB 2nd ICS loud, harsh, systolic	Present	Inc. RBBB	RV +++ PA ++ Pul. vasc. engorgement	RA	2.1	92/10	1.5 cm.	Died during surgery	Stahlman
4	4 yrs.	M	+	LSB 5th ICS loud, harsh, systolic	Present	RVH	RV +++ RA +++ PA ++ Pul. vasc. engorgement	RA	1.8	65/0	1 cm. ×2 cm.	Died during surgery	Stahlman
5	15 yrs.	F	+++	LSB 3rd ICS loud, rough, systolic	Absent	Probable RVH with S wave in all leads	RV ++ RA ++ Pul. vessels normal	RA	1.6	45/7	1 cm.	Successful closure	Kirby
6	31 yrs.	F	+++	LSB 4th ICS loud, harsh, systolic	Present	Complete RBBB	RV ++ RA ++ LV 0 LA 0 PA ++ Pul. vasc. markings inc.	RA	—	/1	1.5 cm.	Died during surgery	Lynch
7	15 yrs.	M	0	LSB 4th ICS loud, holosys.	Present	Inc. RBBB	RV + PA +	RA RV ?	1.8 1.1	25/5	5 mm.	Successful closure	Gerbode
8	11 yrs.	F	+++	LSB 4th ICS loud, holosys.	Present	Delayed intrinsicoid deflection RV	RV ++ LA 0 RA ++ LV 0 PA ++ Aorta small Pul. vasc. congestion	RA	1.7	41/1	12 mm. × 7 mm.	Successful closure	Gerbode
9	5 yrs.	M	++++	LSB 4th ICS loud, holosys.	Present	Delayed intrinsicoid deflection RV P pulmonale P-R = 0.20 sec.	RA ++ LA ++ RV +++ LV ++ Aorta small. Pul. vasc. engorgement	RA RV Cath. entered LV	3.5	60/0	2 cm.	Successful closure	Gerbode
10	42 yrs.	M	++++	LSB 5th ICS loud, holosys.	Present	Atrial fibrillation. Abnormal T waves	RV +++ RA ++ LA ++ PA ++ Pul. vasc. engorgement & hilar dance	RA	1.8	51/9	1.5 cm.	Successful closure	Gerbode
11	17 yrs.	F	++	LSB 4th ICS loud, holosys.	Present	Borderline record P-R = 0.21 sec.	RV ++ PA prom. Hilar dance	RV	2.0	44/4	12 mm.	Successful closure	Gerbode

Cardiac catheterization studies indicate a left-to-right shunt at the atrial level and occasionally evidence of an additional shunt at the ventricular level. Pulmonary hypertension is rarely severe and a right-to-left shunt is absent.

A combination of an atrial septal defect and a ventricular septal defect of the usual variety may of course present most of the features described above. Two methods, however, may aid in excluding the possibility of an atrial septal defect of the secundum variety. If the catheter is inserted into the saphenous vein the tip can frequently be advanced through the foramen ovale into the left atrium. If only a left ventricular-right atrial shunt is present the left atrial pressure will be higher than the right-atrial pressure by 5 mm. Hg or more. If an atrial septal defect is present these pressures will be equal unless the defect is small.[3] Use of the phonocatheter has demonstrated that in atrial septal defect the murmur is maximal at the pulmonary valve, but is faint in the right ventricle and atrium.[12, 13] In the presence of a left ventricular-right atrial shunt the murmur should be maximal in the right atrium since this was the location of the thrill palpable at surgery in the patients described in this study.

On the basis of these features it should be possible to suspect the diagnosis of this distinctive lesion in a majority of patients. This is particularly important for two reasons: (1) An erroneous diagnosis of an atrial septal defect may lead to an unsuccessful attempt to repair the defect using a closed technique. This occurred in Case 2. Correct preoperative diagnosis will indicate open repair of the lesion employing extracorporeal circulation, which is the procedure of choice. (2) The severe symptoms and the clinical picture may suggest the presence of a complicated lesion with a poor prognosis such as an A-V communis with congestive failure and surgery may not be attempted. Repair of the lesion, however, is relatively simple, using the open technique through an incision in the right atrium, and the functional results are excellent.

Summary

Five patients have had successful closure of a shunt between the left ventricle and right atrium with the aid of extracorporeal circulation. The lesion consists of a high ventricular septal defect associated with a defect of the septal leaflet of the triscuspid valve which allows left ventricular blood to enter the right atrium. Characteristic clinical and physiologic features are present which should permit an accurate preoperative diagnosis and thereby lead to proper surgical correction.

Upon temporary manual closure of the shunt bradycardia and a rise in systemic blood pressure occurred. This is apparently the same phenomenon observed by Nicoladoni [17] and Branham [2] as occurring when an arteriovenous fistula is closed.

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DR. DENTON A. COOLEY: In our experience with open repair of septal defects we have encountered six examples of left ventricular to right atrial shunt—a lesion which we call ventriculo-atrial defect. All of these were repaired through a right atriotomy. Previously we suggested that the transatrial approach to ventricular defects of the membranous septum would be a useful method of repair of these defects. Experience has subsequently revealed that the transatrial approach is only suitable for ventriculo-atrial defects and for Type III ventricular defects. In the latter type the ventricular communication lies beneath the posterior leaflet of the tricuspid valve and may be difficult to approach through the ventricle without dividing papillary muscles and chordae tendinae. Through the atrium it may be possible to expose the defect using a longitudinal incision in the tricuspid leaflet.

In most of our patients with ventriculo-atrial defects there was some defect or cleft of the septal leaflet of the tricuspid valve. One patient had a diverticulum of the tricuspid valve as if the leaflet had been split into two layers by the jet-like stream of blood from the left ventricle.

I have enjoyed the splendid presentation which Dr. Melrose has made and congratulate the other authors.

DR. DENNIS MELROSE (closing): Thank you, Dr. Cooley.

I have little to add. I feel the most successful aspect of the paper is its help to the cardiologist. We have noticed that on occasion the internist who has done the cardiac catheterization is so much at variance with his colleagues who have examined the patient only by auscultation that even acrimony is possible. Perhaps the recognition of this distinct clinical picture may help prevent this.